

**Amendments to the Claims:**

This listing of claims will replace all prior versions, and listings, of claims in the application:

**Listing of Claims:**

Claims 1-22 (cancelled).

Claim 23. (new) A genetically transformed plant able to produce a lysosomal enzyme of animal or human origin, said plant transformed via the use of an expression vector comprising:

- a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;
- b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for the expression of the lysosomal enzyme in active form; and
- c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;

wherein said lysosomal enzyme is expressed in seed storage tissues in enzymatically active form and in an amount of at least the 0.8% of the total proteins of the seed.

Claim 24. (new) The plant according to claim 23, wherein the expression vector is a plasmid.

Claim 25. (new) The plant according to claim 23, wherein the promoter is a 7S soy globulin gene promoter or a functional equivalent.

Claim 26. (new) The plant according to claim 23, wherein the signal sequence is a 7S soy globulin signal sequence, or a functional equivalent, and is fused to the sequence encoding the structural portion of the mature lysosomal enzyme deleted of the native signal sequence.

Claim 27. (new) The plant according to claim 25, wherein the signal sequence is a 7S soy globulin signal sequence, or a functional equivalent, and is fused to the sequence encoding the structural portion of the mature lysosomal enzyme deleted of the native signal sequence.

Claim 28. (new) The plant according to claim 23, wherein the lysosomal enzyme expressed in enzymatically active form in seed storage tissues is selected from the group consisting of  $\alpha$ -N-acetylgalactosaminidase, acid lipase, aryl sulfatase A, aspartylglycosaminidase, ceramidase,  $\alpha$ -fucosidase,  $\alpha$ -galactosidase A,  $\beta$ -galactosidase, galactosylceramidase, glucocerebrosidase,  $\alpha$ -glucosidase,  $\beta$ -glucuronidase, heparin N-sulfatase,  $\beta$ -hexosaminidase, iduronate sulfatase,  $\alpha$ -L-iduronidase,  $\alpha$ -mannosidase,  $\beta$ -mannosidase, sialidase, and sphingomyelinase.

Claim 29. (new) The plant according to claim 27, wherein the lysosomal enzyme expressed in enzymatically active form in seed storage tissues is selected from the group consisting of  $\alpha$ -N-acetylgalactosaminidase, acid lipase, aryl sulfatase A, aspartylglycosaminidase, ceramidase,  $\alpha$ -fucosidase,  $\alpha$ -galactosidase A,  $\beta$ -galactosidase, galactosylceramidase, glucocerebrosidase,  $\alpha$ -glucosidase,  $\beta$ -glucuronidase, heparin N-sulfatase,  $\beta$ -hexosaminidase, iduronate sulfatase,  $\alpha$ -L-iduronidase,  $\alpha$ -mannosidase,  $\beta$ -mannosidase, sialidase, and sphingomyelinase.

Claim 30. (new) The plant according to claim 23, wherein said plant is a *Leguminosa*, a cereal, or tobacco.

Claim 31. (new) The plant according to claim 29, wherein said plant is a *Leguminosa*, a cereal, or tobacco.

Claim 32. (new) A method for producing a genetically transformed plant able to produce a lysosomal enzyme, said enzyme being expressed in seed storage tissues of said plant in an enzymatically active form and in an amount of at least the 0.8% of the total proteins of the seed, comprising the following steps:

- constructing an expression vector comprising:
  - a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;
  - b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for expression of the enzyme in

active form; and

- c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;
- transforming plant cells with said vector; and
- using said cells to regenerate said transformed plant.

Claim 33. (new) The method according to claim 32, wherein said plant is a *Leguminosa*, a cereal, or tobacco.

Claim 34. (new) A seed of genetically modified plant able to express a lysosomal enzyme, wherein:

- said seed contains an expression vector comprising:
  - a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;
  - b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for the expression of the enzyme in active form; and
  - c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;
- said enzyme is contained in seed storage tissues in enzymatically active form and in the amount of at least the 0.8% of the seed total proteins.

Claim 35. (new) The seed according to claim 34, wherein the expression vector is a plasmid.

Claim 36. (new) The seed according to claim 34, wherein the promoter is a 7S soy globulin gene promoter or a functional equivalent.

Claim 37. (new) The seed according to claim 34, wherein the signal sequence is a 7S soy globulin signal sequence, or a functional equivalent, and is fused to the sequence encoding the structural portion of the mature lysosomal enzyme deleted of the native signal sequence.

Claim 38. (new) The seed according to claim 36, wherein the signal sequence is a 7S soy globulin signal sequence, or a functional equivalent, and is fused to the sequence encoding the structural portion of the mature lysosomal enzyme deleted of the native signal sequence

Claim 39. (new) The seed according to claim 34, wherein the lysosomal enzyme expressed in enzymatically active form in seed storage tissues is selected from the group consisting of:

$\alpha$ -N-acetylgalactosaminidase, acid lipase, aryl sulfatase A, aspartylglycosaminidase, ceramidase,  $\alpha$ -fucosidase,  $\alpha$ -galactosidase A,  $\beta$ -galactosidase, galactosylceramidase, glucocerebrosidase,  $\alpha$ -glucosidase,  $\beta$ -glucuronidase, heparin N-sulfatase,  $\beta$ -hexosaminidase, iduronate sulfatase,  $\alpha$ -L-iduronidase,  $\alpha$ -mannosidase,  $\beta$ -mannosidase, sialidase, and sphingomyelinase.

Claim 40. (new) The seed according to claim 38, wherein the lysosomal enzyme expressed in enzymatically active form in seed storage tissues is selected from the group consisting of  $\alpha$ -N-acetylgalactosaminidase, acid lipase, aryl sulfatase A, aspartylglycosaminidase, ceramidase,  $\alpha$ -fucosidase,  $\alpha$ -galactosidase A,  $\beta$ -galactosidase, galactosylceramidase, glucocerebrosidase,  $\alpha$ -glucosidase,  $\beta$ -glucuronidase, heparin N-sulfatase,  $\beta$ -hexosaminidase, iduronate sulfatase,  $\alpha$ -L-iduronidase,  $\alpha$ -mannosidase,  $\beta$ -mannosidase, sialidase, and sphingomyelinase.

Claim 41. (new) The seed according to claim 34, wherein said seed is of a *Leguminosa*, a cereal or tobacco.

Claim 42. (new) The seed according to claim 40, wherein said seed is of a *Leguminosa*, a cereal or tobacco.

Claim 43. (new) A method for producing a seed of genetically modified plant able to express a lysosomal enzyme, said enzyme being contained in said seed storage tissues in an enzymatically active form and in the amount of at least the 0.8% of said seed total proteins, comprising the following steps:

- constructing an expression vector comprising:

- a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;

- b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide

the post-translational modifications required for expression of the enzyme in active form; and

c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence; and

- transforming plant cells with said vector; and- using said cells to regenerate transformed plants able to produce said seeds.

Claim 44. (new) The method according to claim 43, wherein said seed is of a *Leguminosa*, a cereal or tobacco.

Claim 45. (new) A method for extracting and purifying the lysosomal enzyme in active form contained in the seed of a genetically modified plant able to express a lysosomal enzyme, said enzyme being contained in said seed storage tissues in an enzymatically active form and in the amount of at least the 0.8% of said seed total proteins,, comprising the following steps:

a. grinding said seed in liquid nitrogen in the presence of an extraction buffer;

b. centrifuging the resulting solution ;

c. recovering and filtering the supernatant with filters having a porosity suitable to the enzyme dimensions; and

d. further purifying the partially purified enzyme by HPLC chromatography.

Claim 46. (new) A method of use of a seed of a genetically modified plant able to express a lysosomal enzyme, said enzyme being contained in said seed storage tissues in an enzymatically active form and in the amount of at least the 0.8% of said seed total proteins, comprising preparing a medicament for enzyme replacement therapy that comprises the lysosomal enzyme extracted from said seed.

Claim 47. (new) The method use of the seed according to claim 46 for the preparation of a medicament for an enzyme replacement therapy in Gaucher disease.

Claim 48. (new) The method of use of the seed according to claim 46 for the preparation of a medicament for an enzyme replacement therapy in Anderson-Fabry disease.

Claim 49. (new) The method of use of the seed according to claim 46 for the preparation of a medicament for an enzyme replacement therapy in Pompe disease.

Claim 50. (new) A method of use of a seed of a genetically modified plant able to express a lysosomal enzyme, comprising the steps of:

- a. transforming a plant with an expression vector of claim 1 to yield the genetically modified plant;
- b. growing the genetically modified plant;
- c. harvesting the seed of the genetically modified plant;
- d. purifying the lysosomal enzyme from the seed; and
- e. preparing a medicament for enzyme replacement therapy that comprises the purified lysosomal enzyme from step "e".

Claim 51. (new) A method of use of a seed of a genetically modified plant able to express a lysosomal enzyme, said enzyme being contained in said seed storage tissues in an enzymatically active form and in the amount of at least the 0.8% of said seed total proteins, comprising and preserving the lysosomal enzyme in enzymatically active form by storing said seed.